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## **Newborn girl with coarctation of the aorta and anomalous left coronary artery from pulmonary artery, with retrograde perfusion of left circumflex artery: a case report**

Hölscher, Martin ; Knirsch, Walter ; Dave, Hitendu ; Burkhardt, Barbara E U

**Abstract:** Background Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare coronary abnormality. Although it exists usually as an isolated abnormality, ALCAPA has been described with aortic pathologies like coarctation or aortopulmonary window. Case summary An 18-day-old female was admitted to the paediatric intensive care unit because of a heart murmur and weak femoral pulses. A transthoracic two-dimensional echocardiography was performed and confirmed suspected diagnosis of aortic coarctation. In addition, a total retrograde perfusion of the left circumflex coronary artery (LCX) was found, without visible flow through the ostium of the left coronary artery (LCA) into the aorta. A coronary angiography was performed, showing a single right coronary artery with a normal right posterior descending artery (RPD). Supplied by collaterals from the RPD, the LCX was perfused retrogradely, passing by the lateral wall of the ascending aorta without flowing into it, but into the right pulmonary artery. At 23 days of age, surgery was performed with resection of the aortic coarctation and reimplantation of the LCA into the posterior aortic wall. Discussion This case demonstrates that coronary artery anomalies like ALCAPA may occur together with other cardiac malformations. Despite concomitant cardiac lesions, careful assessment of the coronary arteries is mandatory, including cardiac catheterization in case of doubt.

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# Newborn girl with coarctation of the aorta and anomalous left coronary artery from pulmonary artery, with retrograde perfusion of left circumflex artery: a case report

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## Background

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare coronary abnormality. Although it exists usually as an isolated abnormality, ALCAPA has been described with aortic pathologies like coarctation or aortopulmonary window.

## Case summary

An 18-day-old female was admitted to the paediatric intensive care unit because of a heart murmur and weak femoral pulses. A transthoracic two-dimensional echocardiography was performed and confirmed suspected diagnosis of aortic coarctation. In addition, a total retrograde perfusion of the left circumflex coronary artery (LCX) was found, without visible flow through the ostium of the left coronary artery (LCA) into the aorta. A coronary angiography was performed, showing a single right coronary artery with a normal right posterior descending artery (RPD). Supplied by collaterals from the RPD, the LCX was perfused retrogradely, passing by the lateral wall of the ascending aorta without flowing into it, but into the right pulmonary artery. At 23 days of age, surgery was performed with resection of the aortic coarctation and reimplantation of the LCA into the posterior aortic wall.

## Discussion

This case demonstrates that coronary artery anomalies like ALCAPA may occur together with other cardiac malformations. Despite concomitant cardiac lesions, careful assessment of the coronary arteries is mandatory, including cardiac catheterization in case of doubt.

## Keywords

Case report • ALCAPA • Coronary transfer • Coarctation of the aorta

## Learning points

- Anomalous left coronary artery from pulmonary artery is a rare coronary abnormality but may occur together with other cardiac malformations like coarctation of the aorta.
- Despite diagnosis of a congenital heart disease like aortic coarctation, careful assessment of coronary arteries is always mandatory.

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Introduction

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare coronary abnormality, occurring in 0.002% of the population.<sup>1</sup> Although it exists usually as an isolated abnormality, ALCAPA has been described with aortic pathologies like coarctation or aortopulmonary window.<sup>2</sup> We describe an interesting case of a newborn girl with coarctation of the aorta and ALCAPA, with retrograde perfusion of the left circumflex artery.

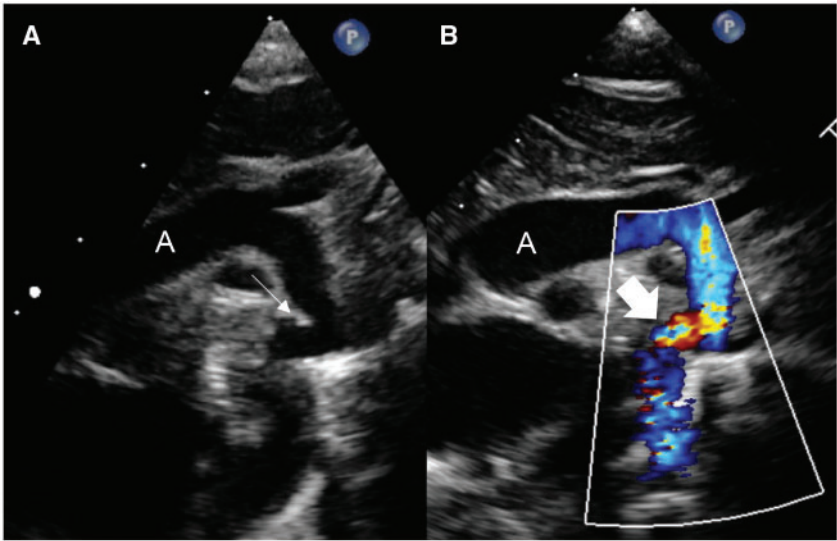
Timeline

Initial presentation	Admitted to paediatric intensive care unit with suspected diagnosis of coarctation. Echocardiography confirmed aortic coarctation and identified coronary anomaly with retrograde perfusion of left circumflex coronary artery.
Day 3	Coronary angiography.
Day 5	Surgery.
Day 12	Post-operative coronary angiography, confirming antegrade perfusion of reimplanted left coronary artery.
Day 23	Discharge home in good clinical condition.
1 month post-procedure: follow-up in cardiologist's office	Feeding and growing well, no signs or symptoms of heart failure.

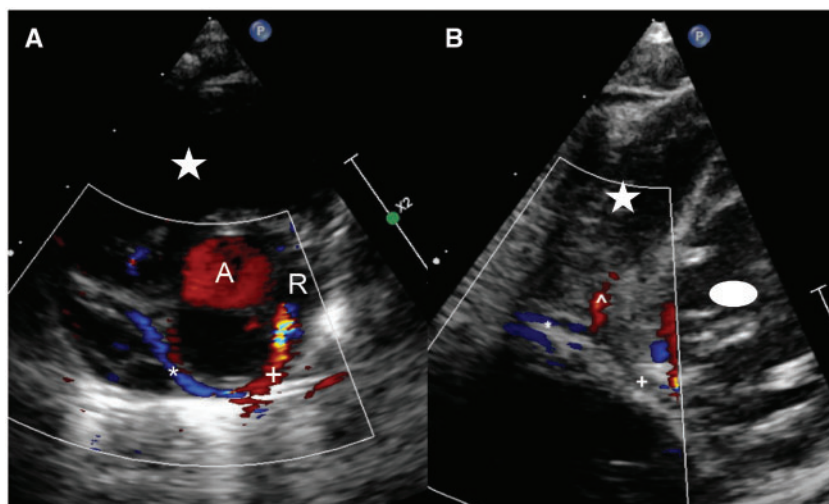
Case report

An 18-day-old female was admitted to the paediatric intensive care unit because of a heart murmur and weak femoral pulses. The patient was in a good clinical condition with spontaneous breathing on room air and normal vital signs, except for a blood pressure gradient of 30 mmHg between the upper and the lower extremities. A trans-thoracic two-dimensional echocardiography was performed and confirmed the suspected diagnosis of aortic coarctation, with a systolic pressure gradient of 30 mmHg and a closed arterial duct (Figure 1). In addition, a total retrograde perfusion of the left circumflex coronary artery (LCX) was found, without visible ostial blood flow at the left aortic sinus (Figure 2, Supplementary material online, Video S1). The left ventricle had a normal ejection fraction without regional wall abnormalities or mitral valve regurgitation. Because of uncertainty regarding the anatomy, a coronary angiography was performed, showing no left main coronary artery, but a single right coronary artery (RCA) arising from the aorta. The LCX was perfused retrogradely via collaterals of a normal right posterior descending artery and it reached the lateral wall of the ascending aorta, not flowing into it but into the right pulmonary artery (RPA) (Figure 3, Supplementary material online, Video S2). Arising from the LCX, small branches of a rudimentary left anterior descending artery were seen. The pulmonary artery pressure was normal.

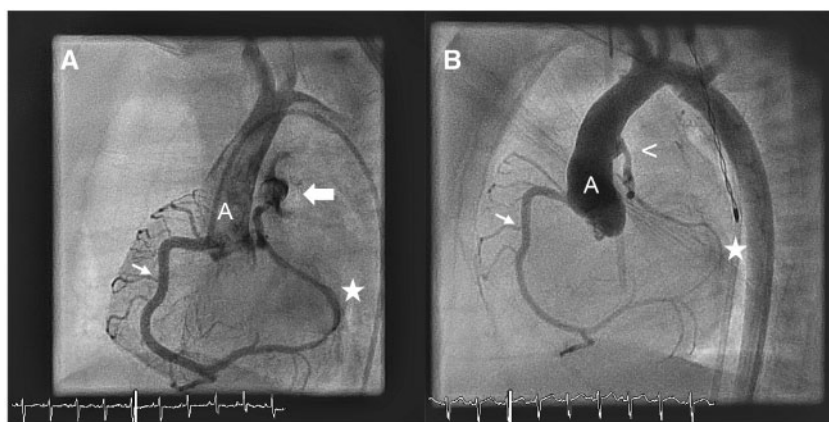
At 23 days of age, surgery was performed with resection of the aortic coarctation, end-to-end anastomosis, and reimplantation of the left coronary artery (LCA) into the posterior aortic sinus, using a button technique without stretching or torsion of the coronary artery (Figure 4, Supplementary material online, Video S3). The RPA defect was closed with a xenopericardial patch. On post-operative Day 4, the patient exhibited recurrent episodes of supraventricular tachycardia, which were successfully treated with flecainide. After surgery, routine coronary angiography was per-



**Figure 1** Coarctation of the aorta. (A) Two-dimensional echo and (B) colour Doppler. A, aorta; \blacktriangledown, aortic coarctation; \blacktriangledown, aortic coarctation with colour Doppler.



**Figure 2** Left circumflex artery arising from right coronary artery. (A) Short-axis view and (B) apical four-chamber view posteriorly. \*, right coronary artery; +, left circumflex coronary artery, Δ, posterior coronary collateral; ★, right ventricle; ●, left ventricle; A, aorta; R, right pulmonary artery.



**Figure 3** (A) Pre-operative angiography of left circumflex artery conducting blood from the right coronary artery, with contrast flowing into right pulmonary artery. (B) Post-operative angiography after reimplantation of the left circumflex artery into the posterior aortic wall. ★, right coronary artery; ★, left circumflex artery; ◀, contrast flowing into right pulmonary artery; <, reimplantation of left circumflex artery into aorta; A, aorta.

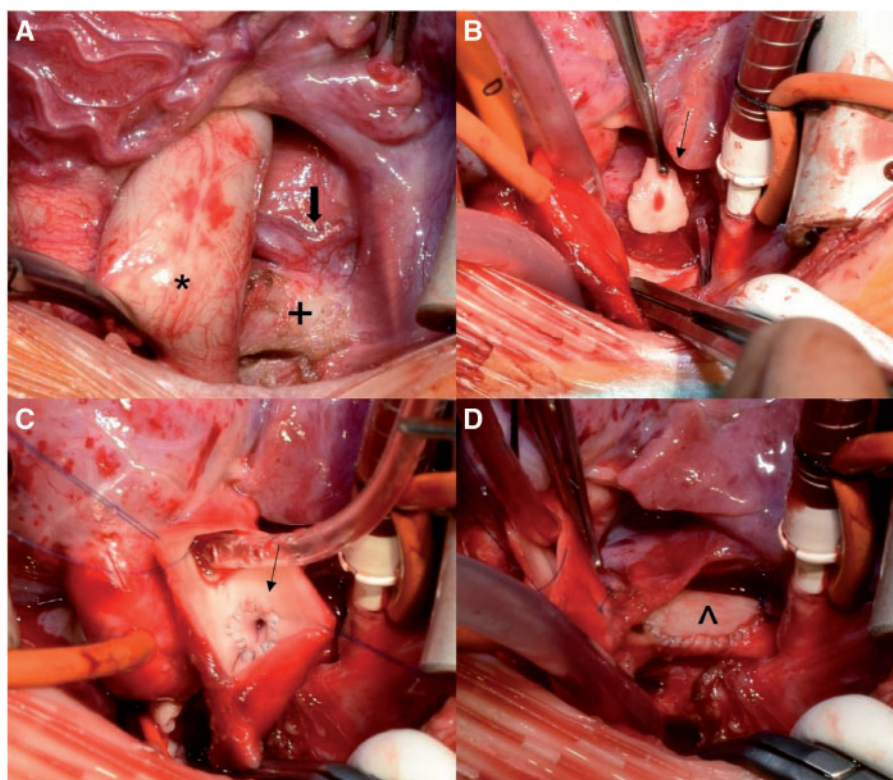
formed, showing normal, antegrade flow into the LCA without stenosis (Figure 3B). The girl recovered well from the operation and was discharged home soon with normal troponin values.

## Discussion

Coronary artery anomalies like ALCAPA may occur together with other cardiac malformations. For example, other aortic pathologies like bicuspid aortic valve or coarctation of the aorta can be found.<sup>3</sup> In our institution, we recently had an additional case with LCX arising from the RPA in a girl with hypoplastic left heart syndrome.

Given the good clinical condition of the patient described in this report, careful assessment with all the available diagnostic tools including cardiac catheterization was warranted and did not cause a harmful delay of surgical correction. Otherwise, immediate catheterization and emergency surgery for reimplantation of the LCA and correction of the concomitant lesion(s) would have been indicated. Intraoperatively, identification of a small aberrant coronary artery may be difficult, but accidental severing of the artery may be life-threatening. In this case, coronary transfer was performed using a button technique in analogy to the button technique used in arterial switch operation.<sup>4</sup> Since coronary obstruction is a known complication of coronary artery transfer, regular, and long-term cardiology





**Figure 4** Surgical correction. (A) Left coronary artery arising from right pulmonary artery. (B) Isolated left coronary artery with button technique. (C) Implanted button with left coronary artery in aorta. (D) Right pulmonary artery defect closed with pericardial patch. \*, aorta; +, right pulmonary artery; ↓, left coronary artery; ↓, button with left coronary artery; ^, pericardial patch.

follow-up will be scheduled for this patient. Anticoagulation was currently not deemed necessary.

## Conclusion

In addition to diagnosis of a congenital heart disease like aortic coarctation, careful assessment of the coronary arteries is always mandatory, as coronary anomalies may occur together with other cardiac lesions. This case shows, in addition, reversed blood flow in the anomalous LCA by echocardiography exceptionally clearly. In case of uncertainty of coronary anatomy or blood flow using two-dimensional echocardiography, cardiac catheterization, and coronary angiography is indispensable before going to surgery to correct the main cardiac defect. If coronary transfer is necessary like in patients with ALCAPA, the button transfer technique can be used together with correction of the concomitant lesions.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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